Biatrial Myxomas With Various Manifestations
– Carney Complex –

Yoichi Yamashita, MD; Taiko Horii, MD

Figure. (A) Preoperative echocardiogram showing 2 large tumors in both atria, almost herniated into the atrioventricular valves. The right-sided tumor is larger in size, $30 \times 35$ mm, than the left-sided tumor, $25 \times 30$ mm. The macroscopic appearance of the 2 tumors is very different. (B) The right atrial myxoma has a multilocular structure. (C) The left atrial myxoma has a villous structure. (D) Loupe image of the left atrial tumor showing villous formation and abundant mucin component. (E) Microscopy of this tumor showing redundant mucinous substrate inside the cells, compatible with myxoma.
A 20-year-old woman was referred to hospital for treatment of large tumors in both atria. On echocardiogram, large myxomas existed in both atria and were nearly herniated into the atrioventricular valves (Figure A; Movie S1). The patient therefore underwent urgent excision of both myxomas using cardiopulmonary bypass through minimally invasive surgery. The right-sided tumor arose from the inferior wall of the right atrium near the inferior vena cava, and the left-sided tumor was attached to the septum. Both tumors were excised with their stalks including the atrial wall, and cryoablation was applied to the surrounding tissue to prevent recurrence. The 2 tumors appeared different macroscopically, but the pathology was the same, and was compatible with myxoma (Figures B, C). With regard to the gross appearance of the cardiac tumors, the right atrial tumor had a multilocular structure compatible with cardiac myxoma, and the left atrial tumor had a villous structure. Microscopy of the left atrial tumor showed redundant mucinous substrate inside the cells, a finding compatible with myxoma (Figures D, E). The postoperative course was unremarkable and the patient was discharged 10 days after surgery. On admission the patient was asymptomatic with slight moon face and sparse facial hair, but she had a long history of multiple surgery for tumors in her whole body. She had a myxoma on the external auditory meatus at the age of 2, multiple myxomas on the vulva, the eyelid, and the auricle at the age of 7, myoma of the uterus at the age of 18, and myxomas on bilateral nipples 6 months before the excision of biatrial myxomas. Given the combination of biatrial myxomas and multiple excisions of myxomatous tumors, the patient underwent thorough endocrinological assessment. She was diagnosed as having acromegaly, Cushing’s syndrome, and hypercoagulable nodule of the thyroid. Positron emission tomography-computed tomography may help rule out any other endocrine overactivity to rule out Carney complex.

Given the combination of biatrial myxomas and multiple excisions of myxomatous tumors, the patient underwent thorough endocrinological assessment. She was diagnosed as having acromegaly, Cushing’s syndrome, and hypercoagulable nodule of the thyroid. Positron emission tomography-computed tomography may help rule out any other inflammatory disease. She underwent endoscopic trans-sphenoidal resection of the pituitary hyperplasia to treat acromegaly 6 months after the open heart surgery, followed by laparoscopic excision of the bilateral adrenal glands, of which the pathological diagnosis was primary pigmented nodular adrenocortical disease. Those findings are compatible with Carney complex. Since then she has been under close observation by the endocrinologist and in a stable condition without any signs of recurrence of cardiac myxoma for more than 6 years.

Atrial myxoma is the most common form of cardiac tumor, and not uncommon in daily practice. Most myxomas arise from the left atrium and multiple occurrence is rare, and that of biatrial myxomas especially so. In the present case, occurrence of biatrial myxomas led to the patient being examined for endocrine disorder, even though she was relatively asymptomatic with only slight moon face and hypertrichosis. Some patients with multiple occurrence of cardiac myxoma or first-time singular cardiac myxoma have been identified as having Carney complex. Carney complex is an autosomal dominant syndrome and was first described by the pathologist J. Aidan Carney. Various manifestations were noted: spotty skin pigmentation in 80% of patients with Carney complex, cardiac myxoma in one-half, cutaneous fibromyxoid tumors in one-third, primary pigmented nodular adrenocortical disease in one-fourth, and acromegaly in one-tenth. It is important that the patient with cardiac myxoma in any form, either solo in the left atrium, or recurrence, or biatrial myxomas as in the present case, be examined for any endocrine overactivity to rule out Carney complex.

The present patient had various manifestations: biatrial myxomas, multiple myxomatous tumors over her whole body, acromegaly, Cushing’s syndrome, and spotty pigmentation of the skin, the most common manifestation, but all other findings were characteristics of Carney complex. Subsequent surgery has maintained the patient as healthy for more than 6 years. Patients with Carney complex have greater potential for tumor recurrence. The primary cause of death in Carney complex is cardiac related, mostly complications of cardiac myxoma, and the patient must be closely followed up.

References

Supplementary Files
Supplementary File 1
Movie S1. Four-chamber view of preoperative echocardiogram. Two large tumors existed in both atria and both tumors are almost herniated into the atrioventricular valves. The right atrial tumor in particular, is moving back and forth through the tricuspid valve.